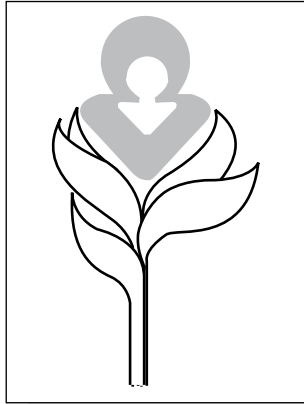


Huntington's Disease Association



Young Person with Juvenile Huntington's Disease at School

Funded by



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Revised 2011

Fact Sheet

A YOUNG PERSON WITH JUVENILE HUNTINGTON'S DISEASE (JHD) AT SCHOOL

What is Juvenile Huntington's Disease?

Huntington's disease (HD) is a hereditary, neurodegenerative condition that causes abnormal movements and other physical signs as well as emotional and cognitive problems. It usually manifests itself in middle life, although it can present at any age. HD gradually worsens with time and there is currently no cure. Juvenile Huntington's Disease (JHD) is the name given to any affected person who has signs or symptoms before the age of 20 years. About 5-10% of all people with HD have Juvenile-onset HD and so it is a rare condition in the general population. This can make dealing with JHD an isolating experience for the young people with HD, their families and also professionals involved with their care. Given their age, they are likely to have regular contact with teachers at school. This leaflet has been written for schools to help them have a better understanding of JHD and hopefully to give them some ideas about what they might be able to do to support a child with JHD. The young people with JHD are likely to be attending a school or college of some kind and as a result will have specific educational and care needs. Some of the needs they may have are discussed below, but it is important to remember that each young person is unique and certain support may, or may not, be needed depending on the young person and their circumstances.

MOVEMENT DISORDER AT SCHOOL

The features of the movement disorder in young people with JHD are complicated and often involve a mixture of rigidity, an irregular staggering gait and myoclonus (brief, strong muscle contractions). The chorea (involuntary movements) often seen in adults with HD is much less common in children with JHD, although it can still appear in some cases.

These movement problems, both rigidity and chorea, affect balance and put the child at a high risk from falls. They will probably find it difficult to get to parts of the school without easy access (e.g., up-stairs rooms). They may also find it difficult to carry their own books and equipment that they need at school and may require assistance. As the condition progresses, it may be necessary for them to be accompanied between classes, especially at times when the corridors are very busy and crowded. It may be useful to think

ahead about what specific parts of the school may cause problems, so that there is time to find funding and establish any changes that need to be made. Often, however, all that is needed to overcome these difficulties is to make a few simple changes (e.g., switching the young person's classes to classrooms with easy access).

COGNITIVE DISORDER IN SCHOOLS

Young people with JHD may also have cognitive (or 'thinking') changes that cause them difficulties in the school environment. In terms of changes to cognition (or 'thinking') in JHD there are a number of things that are fairly common in young people with JHD. For example, they may have a slowed response time and/or a lessened ability to take things in. The problems they have in taking information in tend to get worse as the condition progresses, but often remain fairly intact in the early stages of the condition, even when their responses may be somewhat slowed. They may also find it harder to concentrate, and can have memory problems. It can be harder for them to initiate actions that they may be capable of doing, which may make them appear to be lethargic and lazy. However, all of this is part of the condition, and is easily helped if they have extra help in getting started in activities. They may be more impulsive and have difficulty waiting for things, which is again a part of the condition. They may also have quite fixed thinking, and find change to routines and coping with different things at the same time difficult to deal with. Finally, these difficulties with thinking tend to cause them to have problems with organisational skills (e.g., having equipment and books for homework, for example). They may also mean that they need extra help in classes. Sometimes, this can be given by a friend or the class teacher, although at other times they may require a Learning Support Assistant in some, or all, of their lessons. More detail about this can be found in Jim Pollard's book, 'Hurry up and Wait', and in the book 'Understanding Behaviour in HD' by Jane Paulsen (see 'Further Information').

CHALLENGING BEHAVIOUR IN SCHOOLS

Challenging behaviours are fairly common in JHD, in particular where symptoms begin when the young person with JHD is in their teens. In some, this can be the main obvious symptom with much fewer movement problems

than is normally seen in HD patients. For some years, it may be uncertain whether their behaviour is due to the Huntington's disease or the environment, particularly as many specialists would be reluctant to give a diagnosis of HD (or to refer the young person for a genetic test) without obvious physical signs of HD. It is important to remember that simply because a child has HD, it does not follow that every other problem is related to it. Changes to behaviour can have many different causes, particularly in children / young adults. For a child with HD the family situation can be disrupted and the school may be the most, and sometimes only, stable environment that the child has.

However, it is equally important to remember that the challenging behaviours could be due to the HD and that they should therefore be managed sensitively. When trying to deal with changes in behaviour both in a child with JHD (and in at-risk children and siblings) it is important to bear in mind the implications that labelling the child (either as difficult, or as affected by HD) may have in any particular context. In some cases, being labelled as 'difficult' may be more stigmatising than having a diagnosis of HD. People may be more sympathetic to a child when they believe there is a medical reason for the child's behaviour, but when there is not, there may be a temptation for people to place the blame for the young person's problems on the family. It is important to bear in mind that a child in an HD family needs special attention if they are showing emotional difficulties regardless of whether or not they have the disease. Whether or not their problems are directly caused by HD, they may have had some very serious issues to deal with indirectly related to HD and staff should equally be sympathetic to this. Finally, although challenging behaviours may appear in some children with JHD, in many children they don't appear at all. Further information can be found in the factsheets 'Challenging Behaviour in JHD' and 'Behaviour Problems' and in 'Understanding Behaviour in HD' (see 'Further Information').

EPILEPSY

Epilepsy can be a problem in young people with JHD, although less so when symptoms begin in the teens. Having to deal with epileptic seizures worries many teachers. However, there are many sources of support that can be used to get more information and advice on epilepsy and there is therefore no reason why it should be an insurmountable problem.

Epilepsy Action (see www.epilepsy.org.uk) and the National Society for

Epilepsy (see www.epilepsynse.org.uk) produce a range of information booklets and leaflets, including advice for teachers (see www.epilepsy.org.uk/info/education/professionals.html). Epilepsy Action also have a free-phone helpline (0808 800 5050), which can provide staff with more specialist advice. In addition, it can be helpful to organise staff training (e.g., from a specialist epilepsy nurse) for members of staff who have contact with the child, as this will give them an opportunity to ask questions about issues not covered in the written information and be given up-to-date and specific advice about their type of epilepsy.

SWALLOWING PROBLEMS

Young people with JHD may also have problems with speech and with swallowing. Adults and older children with HD need a high calorie intake. The reason for this at present is unclear as it is not thought that this problem is wholly caused by the involuntary movements often seen in HD. However, it is important that people with HD receive the correct number of calories as it can affect their physical and mental well-being. Given the problems that people with JHD often have with swallowing, and that they may not be able to go home to their parents/guardians at lunchtime, it is important that they are given help to get as many of these calories as possible. To be able to get enough calories, they may need to eat small, frequent snacks in-between meals. They may also need more time to eat, because they will have to eat slowly, and will need a calm, quiet environment as they need to concentrate on eating. If drinking is difficult because of swallowing problems, care must be taken to ensure that they drink enough to avoid getting dehydrated, especially in the summer months. One of the problems often encountered by young people with JHD is that they may become very tired during the school day, and ensuring that they receive adequate calories can help this. Further information can be found in HDA factsheets (see 'Further Information').

SPEECH PROBLEMS IN SCHOOL

Speech problems usually start off with only mild slurring, but this will continue to become increasingly more affected as the condition progresses. Speech and communication problems are one of the aspects of the condition that can make it most difficult for the child with JHD to maintain good relationships

with peers. In class, the young person may need more time to speak, receive the message and reply. Speech aids can be of help, but it is important that an assessment is made early on by a speech and language therapist and that any suggested solutions (e.g., flash cards, computer aids) are put in place as early as possible to allow the young person to get used to using them. It is also important that the school is aware of what aids are being used at home, so that a consistent method is being used. There are a number of simple things you can do to help communication with the child. Further information can be found in HDA factsheets (see 'Further Information').

GENERAL POINTS ABOUT JHD IN SCHOOLS

A number of general comments can be made about the support needs of a young person with JHD. Firstly, it is very important that these support systems that have been discussed are put in place as early as possible. An assessment of the young person's needs should be made early to identify any difficulties they are having, and it must be remembered that these needs are likely to change as the condition progresses and so regular re-assessment is important. The earlier changes are made, the more time the person will have to adapt to these changes, meaning that they are likely to be more useful for longer. For example, if a computer aid is given earlier, the young person will have time to learn how to use it so that they are more likely to be able to use it as their cognitive abilities decline. Also, these changes should be implemented quickly. Having a progressive condition, the needs of a person with JHD changes and a long delay in meeting their needs can mean that the person can no longer get the best benefit from the support that was needed at the time a request was made. However, considering what a young person might need in the future (which will involve assessing how their condition will progress in the future) should be done sensitively and with an awareness of how discussions about the future may affect them and their family.

Although a young person with JHD may progress quickly from the onset of their condition, it is important to remember that this can be very variable and a person with JHD can live a full life for many years. It has known for people with JHD to live for a long time after the onset of their condition. The engagement involved in school activities, the process of learning and the social environment of the school may also play a very important role in the progression of the condition and may help to maintain the current abilities of the person for as long as possible, as well as helping them to develop new ones. It is therefore

very important to support the young person so that they are able to remain at school if they wish to do so as long as possible.

THE FAMILY AND SIBLINGS

The child with JHD has been discussed at length, but for a school it is also important to consider the wider family. Huntington's disease is a condition caused by a dominant gene. This means that when one parent has Huntington's disease, their biological children have a 50% risk of developing the condition. It is therefore important to remember that any siblings, who may be at the same school as the affected child, may be at-risk and have concerns about that. Even if they are not at-risk (e.g., they are not a biological sibling or they already know their genetic status), they still have to cope with a sibling and/or parent who has HD. They may be being bullied (this may also be true for the young child with JHD), or they may feel a responsibility to look after their brother or sister. It is important therefore that the school is also aware of how JHD in the family may be impacting on them and their education and development. It can be very useful for staff, and perhaps also students, at the school to gain a basic knowledge of HD and the genetics of HD as misunderstandings and misinformation can cause many problems.

THE SUPPORTED SCHOOL

With knowledge about HD, support and advice on how to cope with the problems of HD in young people, it can be possible to keep the child at school for a long time. Although these young people may have additional needs, with support there are few things that they cannot do. Schools are in a unique position in being able to support the child and enable them to achieve. Equally, seeing how the young person copes with a progressively disabling condition can be of untold benefit to everyone in the school, both children and adults.

If a school has further questions about JHD, they can contact the Huntington's Disease Association or their Regional Genetics Service / Specialist Huntington's disease clinic. The Huntington's Disease Association has a team of Regional Care Advisers who can offer them support and advice, and answer any questions they may have.

FURTHER INFORMATION

Quarrell, O. et al (2009) *'Juvenile Huntington's Disease and other Trinucleotide Repeat Disorders'* published by Oxford University Press ISBN 978-0-19-923612-1

Quarrell, O. (2008) *'Huntington's Disease: The Facts (2nd Edition)'*, available from the Huntington's Disease Association, www.hda.org.uk

Glendinning, N *'Huntington's Disease in Children and Teenagers: A Guide for Professionals'*, available from the Huntington's Disease Association (www.hda.org.uk).

Jim Pollard, *'Hurry Up and Wait'*, available from www.lulu.com/content/2517713

Jane Paulsen, *'Understanding Behaviour in Huntington's Disease'*, available from the Huntington's Disease Association (www.hda.org.uk).

Huntington's Disease Association factsheets *'Challenging Behaviour in Juvenile Huntington's Disease'*, *'A brief guide to Juvenile Huntington's Disease for Children's Hospices and palliative care services'*, *'Talking to Children about Huntington's Disease'*, *'Information for Teenagers'*, *'Communication Skills'*, *'Behavioural Problems'*, *'Eating and Swallowing Difficulties'*, *'Huntington's Disease and Diet'* available to download from www.hda.org.uk

A Guide to Huntington's Disease for General Practitioners and the Primary Health Care Team, available from the HDA, www.hda.org.uk

Nance, MA (2007) *'The Juvenile HD Handbook: A Guide for Families and Caregivers'*, available from:
www.hdsa.org/images/content/1/1/11702.pdf

'Juvenile Huntington's Disease and the School Experience: Education and the Child Affected by Juvenile Onset HD', available from the Huntington's Disease Society of America (HDSA), www.hdsa.org

Huntington Society of Canada (2000) *'Juvenile Huntington Disease: A Resource for Families, Health Professionals and Caregivers'*, available from:
www.huntingtonsociety.ca/english/uploads/Juvenile_HD_2008.pdf

Huntington's Disease Association 'Huntington's Disease in the Family', available from the HDA, www.hda.org.uk

Gray, A. (2000) 'Huntington's and Me: A Guide for Young People', available from the HDA, www.hda.org.uk

Hennig, B (2003) 'Talking to Kids about Huntington's Disease', available through www.talkingtokidsabouthd.com

FREQUENTLY ASKED QUESTIONS

How long can we expect the young person to live after they develop symptoms?

Unfortunately, there is no answer to this question. Huntington's disease in adults usually evolves over 15-20 years and may run a slightly faster course in younger people. However, how long people live for after they develop HD symptoms is very variable. At the moment, it is very difficult to predict this.

How long can we expect the young person to be with us at the school?

Again, this is very variable. Some young people do very well and manage to stay in a school for a long time, even in a mainstream school. It depends largely on their individual symptoms, the support at the school (both staff and students) and the facilities at the school.

Is a mainstream or special school better for the young person?

More detailed information about this can be found in the book 'Huntington's Disease in Children and Teenagers: A Guide for Professionals,' by Neil Glendinning (see 'Further Information'). However, influencing factors are: (i) The type and degree of disability, (ii) The availability of schools locally, (iii) The response of schools to the problems posed by the disease, (iv) Schools that will meet the physical and emotional needs of the child, (v) the opinions of the parents and the affected youngster (vi) the opinions of the siblings and (vii) the

opinions of the main caregivers and professional advisers. Home schooling is not generally recommended, as the parent is placed under additional pressure and the child is deprived of social contact. Mainstream schooling can become difficult as the disease progresses, but this very much depends on the particular situation. Moving to a special needs school can be a big step for everyone. Therefore, careful thought needs to be given about the timing of such move. A school for children with physical disabilities may be more appropriate than a school for children with severe learning disabilities.

We are having real problems managing challenging behaviour. Is there anything we can do?

The school is torn between being sympathetic to the child with HD and the welfare of other students at the school, and so this can be a really difficult situation. However, there is no simple answer as challenging behaviours could be caused by so many different things and take many different forms. The problems could be due to the HD, or they could be due to a disrupted environment, or both. It may be worth consulting an educational psychologist or a paediatric psychiatrist who knows something about HD. For example, it could be that certain drugs could help, or drugs that are being taken need to be changed (they could need to be stopped, reduced or increased). Also, there may be some behavioural modification techniques that could be tried. Using these two methods together has sometimes had some success. However, it should be remembered that challenging behaviour in HD can sometimes be caused by very small problems that can be resolved quite easily. Is the child finding the busy school environment too overwhelming? Are they having difficulty expressing what they want?

No-one seems to know much about Juvenile Huntington's Disease. Where can I go for help?

Being relatively rare, it can sometimes be difficult to find someone to talk to who knows about Juvenile Huntington's Disease. Probably the best sources of support in these situations are the Huntington's Disease Association, the Regional Genetics Service or Specialist HD clinic. The Huntington's Disease Association have a team of Regional Care Advisers, who can offer advice and training on HD.

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Fact sheets available from the HDA:

- General Information about Huntington's Disease and the HDA
- Predictive Testing for Huntington's Disease
- Talking to Children about Huntington's Disease
- Information for Teenagers
- Eating and Swallowing Difficulties
- Huntington's Disease and Diet
- The Importance of Dental Care
- Communication Skills
- Behavioural Problems
- Sexual Problems
- Huntington's Disease and the Law
- Huntington's Disease and Driving
- Advice on Life Assurance, Pensions, Mortgages etc
- Seating, Equipment and Adaptations
- Checklist for Choosing a Care Home
- Advance Decision to Refuse Treatment (ADRT)
- A Carer's Guide
- Challenging Behaviour in Juvenile Huntington's Disease
- A Brief Guide to Juvenile Huntington's Disease for Children's Hospices and Palliative Care Service
- A Young Person with Juvenile Huntington's Disease at School

All Fact sheets can be downloaded for free from our website:
www.hda.org.uk or ordered direct from Head Office

For a publication price list/order form, membership form, details of our Regional Care Advisers and local Branches and Groups, please telephone, email or write to:

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